

Reticulum cell Sarcoma of the choroid and brain

(RETICULOSARCOME DE LA CHOROÏDE ET DU CERVEAU)

THESE

présentée à la Faculté de médecine de l'Université de Genève
pour obtenir le grade de docteur en médecine

par

Glenn W. OWENS

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Thèse N° 3414

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La Faculté de médecine, sur le préavis du professeur Jean Babel, autorise l'impression de la présente thèse, sans prétendre par là émettre d'opinion sur les propositions qui y sont énoncées.

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Le doyen :
William Geisendorf

Thèse No 3414



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Summary

A case of reticulum cell sarcoma of the eye and brain is reported in a 72 year old man presenting clinically with a unilateral uveitis. A cerebral tumor was diagnosed as reticulum cell sarcoma one year after the onset of ocular symptoms. The cerebral tumor was treated with radiation and the ocular tumor with enucleation. This case has a similar evolution to previously reported cases.

Key words

Uveitis, reticulum cell sarcoma of the eye and brain.

Uveitis associated with reticulum cell sarcoma of the choroid and brain is the subject of this paper. Many cases of various lymphomatous involvement of the brain and meninges, the choroid, the retina, the orbit, and the external eye have been previously been reported.

Bailey (1929) is generally noted by most authors as being the first to describe sarcomas of the brain. These possibly arose from melanotic cells which migrated into the pia-arachnoid and were similar to melanocytes in the choroid coat of the eye. This work was followed by those of Abbot (1943), Albites (1963), Sugarbaker (1940) and Yuile (1938). These authors described lymphomatous involvement of the brain and the meninges. Only Sugarbaker described a case of orbital lymphomatous involvement in a series of 196 cases of which 184 were reticulum cell sarcomas.

Hartshone in 1922 described a case of a 3 year old boy who died from what was thought to be metastatic brain disease which originated in the eye. This case presented as a conjunctivitis and progressed to a nongranulomatous uveitis. He postulated the idea that collections of lymphocytes may exist in the ciliary body and then develop abnormally into tumors.

Reese, Blody and Fredrick in 1954 reported the ocular foci of extramedullary or extrahematopoietic tissue were found in 14 % of autopsied stillborn and new-born infants. Reese in 1963 reported 3 cases of reticulum cell sarcoma of the uvea, one case which presented as a posterior, non-granulomatous uveitis resistant to treatment. He postulated that fundus changes may be due to reactivation of hematopoietic properties of the uvea rather than to a simple infiltration.

Other cases presented by Currey et al. and Neault et al. of the choroid without cerebral involvement. These cases presented as solitary tumors of the choroid associated with a uveitis. Vogel et al. in 1968 reviewed 6 cases of reticulum cell sarcoma of the retina and uvea. Two cases revealed tumors of the same cell type in both the ocular and cerebral tumors; all 6 cases involved the retina; 3 cases presented as non-granulomatous, posterior uveitis. This review was concluded with a statement that there is a suggestion that the reticulum cell sarcoma arises primarily in the eye.

Neault et al. (1973) presented 17 cases of reticulum cell sarcoma of the brain. Seven cases had an associated uveitis. In each case ocular symptoms or uveitis presented before the brain lesions were symptomatic. Uveitis was not related to any other type of intracranial lymphoma. There were no pathologic diagnoses of the ocular conditions given. The uveitis was thought to be a secondary inflammatory process.

Arnesen and Naumann presented each a case of reticulum cell sarcoma of the retina and brain at the European Ophthalmologic Pathology Society meeting, May 1973. These cases had similar histories of posterior, non-granulomatous uveitis, unresponsive to medical treatment for as long as eight months. The neurologic symptoms associated with the brain tumors appeared after the ocular problems had begun in these cases. Each case had a definite pathologic diagnosis of both brain and ocular lesion.

Beasly in 1961 described a case of lymphosarcoma of the choroid which presented a uveitis with absolute glaucoma. The patient was treated with enucleation and radiation, and there was no recurrence reported at the time of writing some three years later.

Hogan (1973), in an oral communication with Babel, has reported two cases of concurrent ocular and cerebral malignant lymphomas treated with enucleation and cerebral radiation. These cases are now stable without evidence of recurrence.

Case report

Patient F. B., a 72 year-old caucasian male, was first seen in the university Eye Clinic Geneva, in May 1969, for a general eye examination. His visual acuity was 1,25 in both eyes; intraocular tension was 16 mm/Hg in the right eye and 18 mm/Hg in the left eye; there were vitreal opacities noted on the vitreo-retinal surface equatorially in the right eye, and degeneration and retinoschisis was noted in the extreme periphery also.

The patient was seen two months later in July 1969 and there were no changes noted.

He next presented to the Eye Clinic, Geneva, two years later, May 1971, because of a diminution of visual acuity in the right eye. Visual acuity was 0,3 in the right eye and 1,25 in the left eye; intraocular tension was 18 mm/Hg in the right eye and 19 mm/Hg in the left eye; there was heavy flare and cells in the vitreous and hemorrhages of the retina of the right eye. Fluorescein angiography of the right eye was considered to have shown a Junius-Kuhnt type of retinal degeneration. The patient was admitted to the hospital and examined for cause of the uveitis. The usual uveitis survey was non-contributory. Electro-retinogram of the right eye was decreased to 100 μ V and was normal; 150 μ V in the left eye. Treatment with 40 mg of Prednisone was effective in controlling the uveitis. By July 1971 the visual acuity had returned to 0.9 in the right eye.

The patient next presented to the clinic November 1971 with the same complaint as the previous visit. Visual acuity in the right eye was light perception. The ocular tension was 17 mmHg. There was an exudative detachment of the retina in the right eye. The vitreous was cloudy centrally with a clear periphery. The electro-retinogram was abolished in the right eye. Echography revealed a serous detachment of the choroid and retina. The left eye was unchanged from the previous examinations. The patient was again treated with topical and systemic steroids, and the vitreous cleared to a state of minimal flare and cells and the exudative detachment of the retina absorbed.

The right eye was subjected to filtering surgery for an episode of glaucoma which was unresponsive to medical treatment during the month of December 1971. Vision at this time was no light perception in the right eye.

The patient was hospitalized April 1972 because of an episode of aphasia, disorientation and a right hemiparesis. He received radiation for a left cerebral hemispheric tumor and subsequently developed a right hemianopsia in the left eye. Examinations for other primary lesions or metastatic were normal. The right eye was noted to be phthisical.

The right eye was enucleated November 1972 because of ocular pain. Diagnosis of choroidal reticulum cell sarcoma was made at this time.

The patient was seen one year later, October 1973. There was no change in the hemianopsia or new neurological loss. Lumbar puncture at this time showed 65 cells which were of the same morphology at those found in the choroidal tumor.

Summary of case

Onset of uveitis - May 1971

Diagnosis of
cerebral tumor - April 1972

Diagnosis of
ocular tumor - November 1972

Pathology

Horizontal sections through the central part of the eye were stained with haematoxylin-eosin and with Gomori's silver method for reticulin.

The choroid is elevated by massive tumor invasion. This invasion is formed of cells which have increased volume, irregular cytoplasm, and a large amount of chromatin. There are numerous mitosis of which some are typical. The cells are inclosed in a network of reticulum fibers. The retina has been completely destroyed. There are many inflammatory cells in area which was the choroid. There is no invasion noted of the sclera.

Discussion

This is a case of well documented reticulum cell sarcoma of the choroid and brain, though it could be argued that the cerebral lesion is not for certain because there has been no direct tissue obtained, only the cells from the cerebral spinal fluid.

This case has an evolution seen in the other previously mentioned. As in most of the other cases, there is no apparent involvement other than the brain and eye. One can note with interest the attempts of those treating the primary ocular processes and the lack of information obtained by the routine uveitis examinations. It is important to note also that cerebral radiation was effective in controlling the cerebral tumor as was the cases that Hogan reported to Babel.

An adequate number of these cases have been reported to be able to consider these tumors as separate but concurrent malignant neoplasms of the central nervous system.

The specific nomenclature given to this type of tumor is varied. Some have referred to it as "microglioma", others "reticulo-endothelial sarcoma", and still others have followed the classification of Rappaport and considered them as perivascular malignant lymphomas, histiocytic type confined to the brain and eye. As future cases are recorded, perhaps a better or one of the above will be accepted as standard classification. Also, with the increased awareness of these tumors, perhaps there will be more complete diagnostic examination made, which one could consider to include lumbar puncture and microscopic examination of vitreous biopsy. An earlier diagnosis of the ocular tumor would certainly reduce the patient morbidity considering the long courses of corticosteroids. There possibly would be a decreased mortality from earlier diagnosis of the cerebral tumor also.

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